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### **Case and Review**

## Long-Term Results of Serial Transverse Enteroplasty with Neovalve Creation for Extreme Short Bowel Syndrome: Report of Two Cases

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### Keywords

Short bowel syndrome  $\cdot$  Citrulline  $\cdot$  Serial transverse enteroplasty  $\cdot$  Ileocecal valve  $\cdot$  Neovalve

### Abstract

**Objective:** The aim of this article was to determine whether serial transverse enteroplasty (STEP) and the creation of a new ileocecal valve in extreme short bowel syndrome (SBS) cases (<45 cm) is effective in intestinal adaptation and improvement of nutritional parameters and serum citrulline levels. **Patients and Methods:** We present 2 cases of SBS treated with STEP. Enterectomy was performed for massive intestinal ischemia secondary to a gastrointestinal stromal tumor in the first case and to catastrophic antiphospholipid syndrome in the second. After enterectomy, the short residual bowel measured 34 cm in the first patient and 45 cm in the second. In both cases STEP, cholecystectomy, and gastrostomy were performed. In the first case a Brooke neovalve was created, and in the other the ileocecal valve was preserved. **Results:** Both patients could finally be weaned off total parenteral nutrition (TPN) and gastrostomy feeding, maintaining a good nutritional status 1 year after surgery. **Conclusions:** In extreme SBS, a minimum length of 80–90 cm of functioning small bowel and an intact ileocecal valve are necessary. We plead for the use of STEP with preservation of the ileocecal valve



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or creation of a neovalve using the Brooke technique in order to achieve the ultimate goal, which is to wean patients off TPN. After a critical review of different surgical techniques, a treatment algorithm is proposed. © 2017 The Author(s) Published by S. Karger AG, Basel

#### Introduction

Short bowel syndrome (SBS) implies anatomical and functional loss of intestine with subsequent absorptive surface deficiency and metabolic alterations [1]. In pediatric groups, mortality rates of 37.5% at 5 years were described [2, 3], due to septic complications (bacterial overgrowth, catheter-related sepsis) [4–6] and total parenteral nutrition (TPN)-related liver failure [7–9]. Intestinal adaptation is a key process through which a patient affected by SBS recovers digestive capacity, depending on the length of the residual bowel, the presence of a ileocecal valve/neovalve, and colon preservation. Other factors that contribute to intestinal adaptation are the individual ability to develop hyperphagia, the increase in and hypertrophy of intestinal villi, the delay of digestive transit and, last but not least, a dedicated multidisciplinary team. The clinical course and serum biomarkers such as citrulline (nonessential amino acid produced exclusively by enterocytes) [10] will guide the therapeutic approach. In case of deficient intestinal adaptation, different surgical options are available. These techniques aim at increasing the absorptive surface through bowel lengthening and restoration of ileocecal valve function in order to prevent bacterial overgrowth. In all cases, the goal is to obtain an adequate nutritional status that can be maintained without TPN [11].

#### **Case Presentation**

#### Case 1

A 52-year-old male underwent massive bowel resection with end jejunostomy and cecostomy without ileocecal valve preservation for mesenteric thrombosis secondary to a gastrointestinal stromal tumor. Gastrointestinal transit showed a residual bowel length of 34 cm (Fig. 1). After 4 weeks of exclusive TPN, citrulline levels were <20  $\mu$ mol/L, and the patient had presented several episodes of dehydration and hyponatremia due to jejunostomy losses. It was decided to restore the intestinal transit with serial transverse enteroplasty (STEP), aimed at obtaining a small bowel length of 80 cm. This required 10 endostaplers of 4.5 cm (calculated length =  $34 \text{ cm} + [10 \times 4.5 \text{ cm}]$ ), placed alternatively onto the antimesenteric and mesenteric border at 3-cm intervals, taking special care to avoid injuring the vascularization (Fig. 2, Fig. 3). A Brooke-type neovalve was created, anastomosing the distal jejunum to the cecum [12, 13]. Cholecystectomy and feeding gastrostomy were also performed. For 5 weeks postoperatively, the patient required TPN and enteral nutrition, oral supplements of pancreatic enzymes, proton pump inhibitors, vitamin B12, and codeine and loperamide as needed. Serum citrulline levels 2 months after surgery surpassed 20 µmol/L. After 2 months, TPN could be stopped and the patient was discharged on an oral specific diet (500 mL) complemented by nocturnal enteral nutrition (1,000 mL), with 2–3 soft stools per day. After 6 months, the gastrostomy was removed. One year after surgery the patient had gained 14 kg (48 vs. 34 kg before surgery) and had a stable nutritional status, with oral supplementation and routine follow-up by a nutritionist.

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### Case 2

A 30-year-old woman developed mesenteric ischemia due to catastrophic antiphospholipid syndrome, requiring a massive intestinal resection with end jejunostomy 45 cm from the Treitz angle and end ileostomy 7 cm from the ileocecal valve. The patient had the 4 criteria of catastrophic antiphospholipid syndrome: involvement of 3 or more organs, simultaneous development of manifestations, antiphospholipid antibodies, and histological confirmation of small vessel thrombosis [14, 15]. In this case, catastrophic antiphospholipid syndrome appeared to be induced by an active tuberculosis that was diagnosed and treated later. Over the next 2 months, the patient required TPN, anticoagulation, and specific treatment for pulmonary tuberculosis reactivation. Constant vomiting was attributed to hyperphagia, a common occurrence related to SBS [9]. Afterwards, she underwent surgery to restore intestinal continuity. Due to necrosis of the terminal ileum with only 4 cm of terminal ileum left, jejunoileal anastomosis at the ileocecal valve was practiced and STEP was performed as in the previous case, using 12 stapler cartridges to obtain a total length of small bowel of 90 cm (calculated length =  $45 \text{ cm} + [12 \times 4.5 \text{ cm}]$ ). Cholecystectomy and gastrostomy were also performed. Postoperatively, the gastrostomy had a persistent output of 1,500 mL/day. Upper gastrointestinal series performed through gastrostomy showed a stop in the third portion of the duodenum. Magnetic resonance angiography confirmed the diagnosis of superior mesenteric artery syndrome with perivascular fibrosis and intravascular thrombus (Fig. 4). Corticosteroid treatment was initiated and a feeding jejunostomy tube was placed endoscopically, with the catheter tip distal to the stenotic area. Subsequent improvement was observed over the next 2 weeks, with a decrease in gastrostomy output and better tolerance of oral liquid nutrition. Citrulline levels before surgery were <20 µmol/L and 2 months after STEP hade risen above this level. TPN was stopped and the patient was discharged with oral supplements. At 1 year after surgery, tuberculosis treatment was completed and intestinal function was correct, with a weight gain of 16 kg (64 vs. 48 kg before surgery). A minimal dose of corticosteroids and low-molecular-weight heparin was necessary as maintenance treatment.

#### Review

SBS is defined as a reduction in the total capacity of the intestine, with or without resection, below the minimum level required for proper digestion and absorption of nutrients as well as to provide an adequate fluid and electrolyte balance [8, 12]. Functional bowel capacity does not depend on its real length, but on its capacity and mucosal integrity. Citrulline is a nonessential amino acid synthesized in the enterocyte from glutamine and other amino acids and is not used for protein synthesis [10, 16]. It is a specific biomarker of intestinal function in patients with normal renal function [11, 17] and a good marker of enterocyte load. Optimal serum levels should be at least 20 µmol/L [18]. If these levels are not reached in due time, surgical techniques of bowel lengthening should be used.

Planning of surgery in cases of SBS should take into account the presence or absence of the ileocecal valve [12–14]. It acts as a barrier against reflux of colonic contents and flora and regulates the flow of fluids and nutrients from the bowel to the colon. If it is preserved, the time of contact between nutrients and intestinal mucosa increases, allowing more absorption to take place. Otherwise, rapid transit and small intestinal bacterial overgrowth (SIBO) weaken the patient's condition.

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Colon preservation is crucial for adaptation. The colon retains fluids and electrolytes, absorbs proteins, and rescues unabsorbed carbohydrates due to bacterial metabolism, transforming them into short-chain fatty acids which are directly absorbed into the portal circulation [15, 17–19]. Additionally, the colon decreases digestive motility and stimulates adaptation through peptide YY release. Nevertheless, there are some drawbacks related to colon preservation in patients with SBS. One of them is the increase in the amount of free oxalate that reaches the colon and which is absorbed and excreted by the kidneys, increasing the incidence of calcium oxalate stones. Another drawback is the generation of D-lactates (through fermentation of unabsorbed carbohydrates) which are absorbed and cause neurological symptoms such as chronic fatigue, confusion, convulsions, and in some cases coma.

One classification distinguishes 3 types of SBS (Fig. 5), depending on the presence or absence of a jejunostomy and the preservation or not of the ileocecal valve and the colon [8]. Other authors subdivide SBS patients into absorbers (if they have >80 cm of jejunum) and secretors (if the length is <80 cm) [19]. Each SBS type has disadvantages: (1) Jejunostomy causes important fluid loss resulting in a lack of water, sodium, and magnesium as well as a volume reduction of digestive secretions (gastric and pancreatic juice and bile). Its consequences are malabsorption, malnutrition, and gallstone formation [8]. (2) Jejunocolic anastomosis causes malabsorption with diarrhea, steatorrhea, malnutrition, vitamin B12 deficiency, and renal and biliary stones.

Intestinal adaptation is a process which attempts to restore the intestinal absorption of nutrients, minerals, vitamins, and water [11, 16]. This process involves 3 factors [19, 20]: hyperphagia, structural adaptation, and functional adaptation. Structural adaptation consists of an increase in diameter and height of the intestinal villi, thickening of the intestinal wall, and increase in endoluminal diameter, with a subsequent increase in absorption surface. Functional adaptation consists of an increase in the rate of nutrient absorption due to structural changes, slower intestinal transit, and molecular events such as a change in enterocyte intrinsic enzymatic activity transport. Adaptation will depend on the bowel length, the presence or absence of the ileocecal valve and colon, the patient's age, and the multidisciplinary teamwork [21]. The adaptive changes enumerated before will progressively increase the bowel's efficiency. During adaptation, specific light nutrients such as glutamine, short-chain fatty acids, unsaturated fat, ornithine, and nucleotides are necessary in order to stimulate the enterocytes to release trophic factors [22]. Gastrointestinal hormones such as GLP-1 also stimulate adaptation, especially in enterocyte cells of the terminal ileum and colon. SIBO occurs when the regulatory mechanisms of the intestinal flora are altered and there is no barrier against colonization with colonic flora [23, 24]. Streptococci, Bacteroides, Escherichia spp., lactobacilli, and others, in addition to structural adaptation, cause SIBO. The consequences negatively affect digestion, nutrient uptake, and bile acid conjugation. Subsequently, fat malabsorption occurs, with steatorrhea and deficit in vitamins A, D, and E, while vitamin K is not altered as it is synthesized by the bacteria themselves. Bloating is a consequence of carbohydrate malabsorption [25]. In the worst case, SIBO causes inflammatory changes in the mucosa, with increased permeability and an increased risk of bacterial translocation and sepsis. An indirect diagnosis of SIBO is obtained using the hydrogen breath test or by determination of serum levels of D-lactic acid. A direct diagnosis is made by determination of bacterial concentrations in duodenal/jejunal aspirate.

Other secondary factors are anastomotic ulcers, chronic bleeding (iron deficiency anemia), hepatic steatosis [26], and TPN-induced toxicity. Bacterial endotoxin activates inflammatory cytokines (TNF), which in turn interfere with the fusion of membrane transporter hepatocytes [27, 28].

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Surgery in the SBS is indicated in patients who fail to thrive without TPN and who have citrulline levels <20  $\mu$ mol/L despite proper nutrition and sufficient time to adapt. If a patient's absorptive capacity does not improve within 6 months and adaptation does not occur, SBS is diagnosed [29]. However, in patients with an intestinal length <40 cm and without ileocecal valve, SBS can be diagnosed from the beginning.

Before using any lengthening technique, other procedures must be performed in order to improve motility, reduce the dilated areas, and prevent SIBO [24, 30, 31]. Surgeons' mission is to: (1) Remove the dysfunctional intestine (pathological dilatations, incomplete obstructions, postischemic stenosis). Plications or taperings, bipartitions in continuity or double-barreled gun (DBG) (Fig. 5), or STEP can be performed. We consider that we should act on the intestinal dilated segments to improve length and not motility, advocating thus for DBG or STEP. (2) Improve length slowing motility through neovalve creation. This entails the use of any surgical technique that reshapes the short bowel, unable to adapt, either by lengthening it or creating new valves to prevent SIBO and improving absorption time (Fig. 5) [32]. SBS surgery represents every actuation in the bowel in order to expand the area of absorption. First, restore the bowel continuity if the patient has an ostomy. Only the fact of anastomosing the jejunum to the remaining intestine increases length and transit time [33]. Figure 5 illustrate the types of procedures employed to lengthen the bowel. Some procedures are isoperistaltic, such as the Bianchi technique (longitudinal elongation) [34] or the Kimura-Soper technique (transverse elongation) [35] with its innovative concept of anastomosing the seromuscular bowel layer to the liver or to the abdominal wall. Our opinion is that although these classical techniques are conceptually attractive, they are difficult and risky, and we advocate for the use of STEP [35].

Gallstones appear in a high percentage of patients with SBS. Their etiology is related to bile stasis and malabsorption. It is therefore recommended to practice cholecystectomy in patients who require extensive intestinal resections [34]. The aim of surgery in SBS is to preserve or create as much intestinal length as possible and restore the intestinal continuity following a treatment algorithm in each case (Fig. 6). Citrulline levels will guide the therapeutic approach, indicating the necessity of surgery and its efficacy. The key is to restore intestinal continuity for its advantages: it allows the distal intestinal mucosa to regain functionality, slows down intestinal transit time, and increases the absorption of liquids. Nevertheless, in the first phase after reconstruction diarrhea occurs, leading to perineal irritation and possible formation of oxalate nephrolithiasis.

In other cases, when surgery can favor a reasonable adaptation, the options are: (1) If the residual intestine length is <40 cm and the ileocecal valve is preserved, it may be feasible to indicate STEP [36-38]. (2) If the residual intestine length is >80 cm without ileocecal valve, it is necessary to create a new valve and evaluate the necessity of STEP. (3) If the residual intestine length is between 40 and 80 cm, there are several options that we describe in the algorithm presented in Figure 6, including a new technique, the DBG, similar to the Bianchi technique, but without anastomotic risk, stenosis, or loss of bowel secondary to ischemia. In our opinion, the technique described by Kimura and Soper and the tapering technique for segmental dilations are too risky. Other techniques, such as reversed intestinal segments [39, 40], electrical pacing [41–43], antiperistaltic segments [44, 45], use of small bowel neomucosa [11, 46], artificial sphincter [47–49], intestinal recirculation, intrinsic myenteric denervation, or colon interposition, have been anecdotally performed, and it is difficult to generalize their results. Interventions such as elongation after intestinal bipartition (longitudinal intestinal lengthening and tailoring) [50–52] or DBG are useful and applicable in cases of long dilated segments. However, we consider STEP [36] the ideal procedure



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because it avoids the loss of mucosal surface capable of adequate absorption and is devoid of anastomotic risk. Its technique allows to calculate the number of stapler cartridges required to achieve the required length in each case. STEP has intrinsic advantages: it is easy to perform, avoids anastomosis, may increase the bowel length to more than double or triple, and may also be used in a secondary procedure. Our opinion is that any surgical approach must be prudent, without risk of loss or deterioration of the physical surface of the intestine, but always with the aim of weaning the patient off TPN, which is absolutely necessary during adaptation, but dangerous in the long run.

In this article, we reviewed the surgical principles that should be applied when approaching a patient with SBS and the procedures currently available. The large number of published anecdotal techniques indicates that none of them is ideal. The low number of patients in each group and the lack of long-term effectiveness results do not allow to draw definitive conclusions. SBS is a devastating condition, and the main therapeutic goal is to increase the intestinal absorptive surface up to the point that will allow weaning off TPN. In the first stages of SBS, TPN is crucial for survival and afterwards continues to be very important during the adaptation process, until surgery can be performed. Based on these principles and on our experience with the cases described above, we propose a treatment algorithm for SBS (Fig. 6). In our view, STEP is a very useful, easy to perform, and reproducible alternative that allows significant intestinal lengthening without anastomotic risk. The proportion of patients with SBS who achieve full functional autonomy is increasing. Intestinal transplantation will be reserved for cases with a bowel length <40 cm and TPN-related colon or liver sequelae [53–55]. Currently, intestinal transplantation has a questionable and difficult to predict long-term outcome when compared with successful intestinal adaptation.

### **Statement of Ethics**

There are no ethical conflicts.

### **Disclosure Statement**

There are no conflicts of interest.

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Fig. 1. Intestinal transit showed a residual bowel length of 34 cm.



Fig. 2. Serial transverse enteroplasty technique.

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Fig. 3. Intestinal transit after serial transverse enteroplasty with 80 cm of bowel length.



**Fig. 4.** Superior mesenteric artery syndrome also known as Wilkie syndrome. The aortomesenteric angle is 25° (<30°).

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**Fig. 5.** Small bowel syndrome types, neovalve (Brooke Ileostomy), and techniques to solve bowel dilatations (tapering, double-barreled gun, and serial transverse enteroplasty).

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**Fig. 6.** Treatment algorithm for small bowel syndrome. Small bowel <40 cm without ileocecal valve and colon: total parenteral nutrition (TPN), bowel and liver transplantation. Small bowel <40 cm with ileocecal valve and colon: serial transverse enteroplasty (STEP). Small bowel 40–80 cm without ileocecal valve and colon: enteral nutrition and TPN, if citrulline <20: STEP. Small bowel 40–80 cm with ileocecal valve and colon: if citrulline <20: STEP. Small bowel >80 cm with ileocecal valve and colon: correct if citrulline >20. Small bowel >80 cm without valve and colon: neovalve. Small bowel >80 cm dilated with colon: STEP or double-barreled gun (DBG).